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Nature of hearing impairments in Turner's syndrome

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THE NATURE OF HEARING IMPAIRMENTS

IN TURNER'S SYNDROME

By

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If the desire to write is not accompanied by actual writing, then the desire is not to write.

--Hugh Prather

To all my friends in Portland who provided the impetus for completion of this project.

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Chapter 1

LITERATURE REVIEW

Turner's syndrome is a term applied to females with an abnormal chromosome composition, most commonly monosomy X, which results in short stature and sexual underdevelopment. There have been many terms describing the disorder, for example, ovarian agenis, gonadal agenesis, gonadal dysgenesis, and gonadal aplasia (Jones, et al., 1966); however, the most generally accepted label is Turner's syndrome, named for one of the original investigators of the condition (Turner, 1938).

Associated with the syndrome are a variety of somatic and chromosomal anomalies, some or all of which may be present in an individual. These anomalies include:

1. Webbed neck.
2. Low hairline.
3. Malformation of the heart and/or kidneys.
4. Pigmented nevi.
5. Lymphedema.
6. Hypoplastic nails.
7. High arched palate.
8. Shieldlike chest.
9. Widely spaced nipples.
10. Auditory and visual deficits.

11. Chromosomal abnormalities other than 45 XO (mosaicism and/ or deletions) (Anderson, et al., 1969; Ferguson-Smith, et al., 1964; Lindsten, 1963; Silver and Dodd, 1957; Szpunar and Rybak, 1968).

Of the cytogenetic and somatic features of Turner's syndrome, a search through the literature reveals that the auditory deficits associated with the disorder have received little attention. Although only a few studies have been completed, sensorineural hearing loss seems to dominate when investigators discuss auditory deficits in Turner's syndrome (Anderson, et al., 1969; Smith, 1976; Szmeja and Kosowicz, 1967; Valkov, et al., 1975). Of these the Anderson, et al. and Lindsten researches seem to be the most complete.

In the study reported by Lindsten (1963), 20 of the 41 (49 percent) Turner's subjects demonstrated pure sensorineural losses, 12 of which had a characteristic basin-shaped threshold curve or dip which generally occurred between .125-4 kHz with a median magnitude of 37 dB hearing level (HL). Anderson, et al. (1969) in a study conducted with 79 Turner's patients, found similar results. Forty-two of the 79 (53 percent) examined had sensorineural losses, 33 of which had the dip. This dip was generally bilaterally symmetrical, located between .250-4 kHz with a median magnitude of 35 dB HL. In addition to air and bone conduction thresholds, these researchers used the stapedius reflex to distinguish between a sensorineural and a conductive impairment as well as to determine whether a sensorineural loss was of a recruiting or nonrecruiting type. Anderson, et al. found that all but one of the subjects with sensorineural losses had impairments that were recruiting.

In sensorineural losses without recruitment, one would expect the acoustic reflex to occur at approximately the same levels as in normal ears, 70-100 dB sensation level (SL) re audiometric threshold (Jerger, 1970). Jerger, et al. (1972) found in their study that 95 percent of normal acoustic reflex thresholds occurred between 70-100 dB HL while 99 percent occurred between 65-100 dB HL. They also found that when the hearing loss exceeded 60 dB HL an acoustic reflex might not be elicited.

In patients suspected of having cochlear lesions with recruitment, the acoustic reflex may be elicited at sensation levels less than 60 dB. In these cases the reflex generally occurs at the same hearing level as for normal hearing persons, no matter the severity of the hearing loss. Wilber[1] found that the reflex for pure tones generally occurred across a range of sensation levels from 15 to 60 dB in sensorineural hearing losses exhibiting recruitment. When the acoustic reflex is less than 60 dB SL with a hearing loss of less than 85 dB HL, Jerger (1970) believes recruitment may be inferred.

It is difficult to draw any conclusion about the acoustic reflex in the Turner's population from the study conducted by Anderson, et al. (1969) because the criteria for eliciting the reflex and determining the presence of recruitment were not discussed in sufficient detail. Additionally, none of the reflex data obtained with their subjects was provided for review.

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[1]L. Wilber, "Acoustic Reflex Measurement--Procedures, Interpretations, and Variables," *Acoustic Impedance and Admittance--the Measurement of Middle Ear Functions*, eds. A. Feldman and L. Wilber (Baltimore: Williams and Wilkins Co., 1976), pp. 197-216.

From the studies reporting audiometric data in Turner's syndrome, conductive and mixed losses appear to occur 10-25 percent and 3-7 percent of the time, respectively (Anderson, et al. 1969; Lindsten, 1963; Szmeja and Kosowicz, 1967). This is lower than the 33.4 percent incidence of hearing impairment secondary to otitis media and/or tympanic membrane perforation that Shimizu[2] found in an elementary school population.

Anderson, et al. (1969) found that 53 of their 79 patients (68 percent) had experienced middle ear infections. Twelve of the 26 (46 percent) with normal hearing had previous middle ear infections as did all 17 patients with conductive and mixed losses. Twenty-four of the 34 (70 percent) with sensorineural losses also had a history of otitis media. The ear infections generally occurred before age 10, but six had chronic infections after that age at which puberty normally begins. Stratton (1965) reported a case of Turner's with attacks of acute bilateral otitis media dating from early childhood. Szpunar and Rybak (1968) found that all 10 of his patients had attacks of acute otitis media, the first attack usually taking place in infancy or early childhood. Like Anderson's group, they also found the attacks lessened after 10 years of age and in the majority of the children chronic otitis ensued. It is interesting to note that, in the Anderson study of the 17 patients with conductive and mixed losses, 71 percent of them were under 20 years old.

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[2]H. Shimizu, "Etiology and Pathology of Hearing Loss in Children," *Pediatric Audiology,* ed. F. Martin (New Jersey: Prentice-Hall, Inc., 1978), p. 26.

A few authors (Anderson, et al., 1969; Lenz and Kaijser in Lindsten and Fraccaro[3]; Szpunar and Rybak, 1968) have postulated that an abnormal orientation of the eustachian tube may be the cause of the increased frequency of middle ear infections. A nonfunctioning eustachian tube could produce the same effect.

A functioning eustachian tube is an integral part of a normally functioning middle ear and a prerequisite for successful medical or surgical intervention of diseased middle ears. The eustachian tube has at least three physiologic functions with respect to the middle ear: (1) *ventilation* of the middle ear to equilibrate air pressure in the middle ear with atmospheric pressure and to replenish the oxygen that has been absorbed, (2) *protection* from nasopharyngeal sound pressure and secretions, and (3) *clearance* into the nasopharynx of secretions produced within the middle ear. If any of these functions do not occur, a hypofunctioning eustachian tube will result which, in turn, can lead to otitis media and more chronic ear diseases such as tympanosclerosis, cholesteatomas or mastoiditis (Bluestone[4]; Shambough[5]; Siedentop, 1977). One would therefore assume that measurement of eustachian tube function would aid in the

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[3]J. Lindsten and M. Fraccaro, "Turner's Syndrome," *Genital Anomalies and Related Subjects,* eds. M. Rashad and W. Morton (Springfield: Charles Thomas, 1969), pp. 396-456.

[4]C. Bluestone, "Eustachian Tube Dysfunction," *Proceedings of the Second National Conference on Otitis Media,* eds. R. Wiet and S. Coulthard (Columbus, Ohio: Ross Laboratories, 1978), pp. 50-58.

[5]G. Shambough, "Complications of Otitis Media," *Proceedings of the Second National Conference on Otitis Media,* eds. R. Wiet and S. Coulthard (Columbus, Ohio: Ross Laboratories, 1978), pp. 48-50.

identification of individuals at risk for middle ear disease in addition to monitoring tubal function in patients under medical treatment for middle ear disease.

No research on the assessment of eustachian tube function in Turner's syndrome has been reported. With the clinical applications of impedance audiometry, however, the techniques to measure tubal function have become available (Bluestone, et al., 1972; Holmquist, 1969; Williams, 1975).

Williams (1975) proposed a method of assessing eustachian tube function based on a principle similar to the Valsalva and Toynbee techniques in that positive and negative middle ear pressures are created and originate on the canal side of the tympanic membrane rather than the nasopharynx side but with little active participation by the patient. The method outlined by Williams, utilizing an otoadmittance meter, follows:

> 1. Obtain a seal. Have the patient swallow once or twice with the air pressure at 0.
>
> 2. Trace the tympanogram using acoustic conductance with a 660 Hz probe frequency.
>
> 3. Increase the pressure to +400 mm H_2O [+3924 Pa][6] and swallow several times.
>
> 4. Retrace the curve.
>
> 5. Return air pressure to the peak pressure point of the original tracing and have the patient swallow 3 or 4 times.
>
> 6. Decrease air pressure to -400 mm H_2O [-3924 Pa] and swallow 3 or 4 times.
>
> 7. Retrace the curve. (Williams, 1979, p. 341.)

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[6]Pascals.

When swallowing is performed under an extreme positive or negative pressure condition, the middle ear pressure status (recorded on a tympanogram) can be seen to change from its initial resting state only if the eustachian tube opens and equalizes normally. When the tympanic membrane is pushed toward the middle ear cavity by positive pressure (+3924 Pa), a mechanical pressure will be exerted against the aural opening of the eustachian tube and air will be forcefully expelled through the tube after swallowing. With the tube closed again after swallowing, and without swallowing before retracing the tympanogram, the amount of air left in the middle ear cavity will be less than before swallowing. When the tympanogram is retraced, there will be a shift in the peak pressure point to a negative direction, reflecting reduced middle air pressure. The opposite is true when the tympanic membrane is under stress from negative pressure (-3924 Pa) (Williams, 1975). Some ears, however, may demonstrate adequate equalization function for positive pressure but fail for negative pressure. Williams feels this may imply a eustachian tube that is not completely normal and an ear that has a tendency to develop negative middle ear pressures.

Seidemann and Givens (1977) questioned the examination of middle ear pressure solely as an indicator of tubal patency. The literature suggests that approximately 196 Pa of pressure change is needed for one to note a change (Bluestone[7]; Williams, 1975). In a study conducted with 51 children, Seidemann and Givens found that only 20 percent of

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[7]C. Bluestone, "Assessment of Eustachian Tube Function," *Handbook of Clinical Impedance Audiometry*, ed. J. Jerger (Dobbs Ferry, New York: American Electromedics, 1975), pp. 147-148.

the ears tested generated a middle ear pressure change of greater than 196 Pa. They then examined pressure change with a less stringent criterion of 49 Pa. Only 66 percent of their population produced a middle ear pressure change greater than or equal to 49 Pa, thus they suggest that pressure change should not be the only indicator of an adequately functioning eustachian tube.

Middle ear measurements involve the assessment of middle ear function as well, which refers to "the efficiency of the transmission of energy through the middle ear cavity" (Seidemann and Givens, 1977, p. 491). Seidemann and Givens developed an equation which compares tympanometric amplitude changes (measurement of middle ear function) without regard to a calibration standard or unit of measure. They found that, in their study, function changes greater than or equal to 5 percent were demonstrated by 79 percent of the population. They concluded that changes in middle ear function greater than or equal to 5 percent and changes in middle ear pressure greater than or equal to 49 Pa could be considered evidence of adequate eustachian tube function.

Seifert, et al. (1979) collected and analyzed pressure and function changes in 26 adults (48 ears) and found similar results. They recommended an induced pressure of -3924 Pa for the eustachian tube function test because that amount of pressure maximized visibility of changes in both pressure and function.

The present study was conducted to further define the characteristics of the hearing losses associated with Turner's syndrome. More specifically, this investigation studies the audiometric configuration of the hearing loss (sensorineural, conductive, mixed, or presence of dips) and the status of the middle ear system through the use of tympanograms, acoustic reflex thresholds, and a eustachian tube function test.

Chapter 2

METHOD

Subjects

Subjects were divided into two groups. Group I consisted of 29 females ranging in age from 9.50 years to 40 years (median age = 14.4 years) diagnosed as having Turner's syndrome. Group II consisted of 11 normal females ranging in age from 10 years to 38 years (median age = 24.3 years). Each of the subjects in Group II had pure tone thresholds of ≤ 15 dB HL (ANSI, 1969), presented normal acoustic conductance and acoustic susceptance tympanograms (ANSI, 1978), and reported a negative history of recent otic diseases.

Instrumentation

Measurements of auditory sensitivity were obtained utilizing a Grason-Stadler model 1704 audiometer with signals routed through TDH-49 headphones mounted on MX-41/AR cushions. A Grason-Stadler model 1720B electroacoustic otoadmittance meter in conjunction with a Houston model 2000 X/Y plotter and a Beltone model 15C audiometer were used to gather the tympanometric and acoustic reflex data. The instruments were calibrated according to ANSI standards and checked daily.

Chapter 3

PROCEDURE

Previous history of ear infection was based upon an interview with a patient and/or parent, the completion of a questionnaire, and a review of earlier hospital records. Hearing thresholds were determined by air and bone conduction audiometry utilizing a modified Hughson-Westlake ascending technique (Carhart and Jerger, 1959) for the following test frequencies: (1) air conduction, .250, .500, 1.0, 2.0, 3.0, 4.0, 6.0, 8.0 kHz, and (2) bone conduction, .250, .500, 1.0, 2.0, 3.0, 4.0 kHz.

Acoustic susceptance (Ba) and acoustic conductance (Ga) tympanograms were measured at 220 and 660 Hz probe tone frequencies in a decreasing pressure direction according to the method described by Feldman and Wilber (1976). Acoustic reflex thresholds were obtained utilizing a 200 Hz probe tone at frequencies of .500, 1.0, 2.0 kHz and a 660 Hz probe tone at frequencies of 1.0, 2.0 kHz. Reflex measurements were made at peak pressure point as determined by tympanometry. The activating signal was delivered to the earphone at levels well below the expected threshold of the stapedius reflex and the intensity of the signal was then increased in 5 dB steps until a needle deflection synchronous with stimulus onset and offset was visually monitored on the balance meter. Stimulus duration was approximately 1-2 seconds. Threshold was defined as the lowest stimulus intensity level at which a stimulus-locked change in the inferred middle ear muscle activity of the ear contralateral to the probe could be visually detected on the meter.

The method used for analyzing eustachian tube function was adapted from a procedure described by Seifert, et al. (1979). A conductance tympanogram tracing (+2943 to -2943 Pa) was used as a baseline for maximum visualization of subsequent pressure and function changes. After the baseline tracing was obtained, ear canal pressure was returned to the peak pressure point and the subject was asked to swallow three times with liquid. This procedure allowed for the normalization of middle ear pressure subsequent to the induced pressure changes. A positive pressure of 3924 Pa was then introduced into the ear canal, the subject was asked to swallow three times with liquid, and the tympanogram was repeated for 660 Hz conductance. After pressure was again normalized in the manner described, the canal pressure was decreased to a -3924 Pa and the procedure repeated, tracing in a positive to negative direction. The complete procedure was then performed on the other ear.

Middle ear function change (FC percent) was calculated with the following equation (Seidemann and Givens, 1977):

$$FC\% = \frac{FD_2 - FD_1}{FD_1} \times 100$$

where FD_1 represents the difference in middle ear function between 0 and + 1962 Pa with respect to the baseline tympanogram

FD_2 represents the difference in middle ear function between 0 and +1962 Pa with respect to the tympanogram following the eustachian tube test.

A functioning eustachian tube was indicated when a pressure change of ≥ 49 Pa and a function change of ≥ 5 percent were obtained.

Chapter 4

RESULTS AND DISCUSSION

Otitis Media

The literature has suggested that patients with Turner's syndrome may have a higher incidence of middle ear infections than the average population (Engel and Forbes, 1965; Silver and Dodd, 1957; Stratton, 1965). The present investigation found that 28 of the 29 subjects (97 percent) with Turner's had one or more middle ear infections for which they received treatment. Information could not be obtained for one subject. The infections generally occurred before the age of three or four years, but eight subjects continued to have middle ear infections after age 11 when puberty normally begins. These results appear consistent with those reported by Paparella and Kuhn[8] who found that, in the United States, the incidence of otitis media in the first six years of life appears to be greater than 90 percent. Eight of the 15 (57 percent) Turner's subjects questioned experienced their first episode at 12 months of age or younger; six of the 15 (43 percent) had their first infection at three years of age

[8] M. Paparella and S. Kuhn, "Otitis Media: Definitions and Terminology," *Proceedings of the Second National Conference on Otitis Media*, eds. R. Wiet and S. Coulthard (Columbus, Ohio: Ross Laboratories, 1978), pp. 2-8.

or older. These figures are consistent with those reported by Howie[9] in which 47 percent of his normal population had their initial episode of otitis media at 0-11 months of age.

Of the 11 subjects in the control group, six (56 percent) had at least one middle ear infection which required treatment (one contracted her first episode at less than 12 months of age). The initial episode of the other five (50 percent) occurred at two years of age or older.

Examination of these results indicates a higher incidence of otitis media in the Turner's group as compared to the control group; however, when one compares the Turner's group to Paparella and Kuhn's[10] subjects, there is no apparent difference. The inconsistency in results between the control group and Paparella and Kuhn's group may in part be due to the large age differences in the two groups. Paparella and Kuhn's group consisted of children six years of age or younger, whereas the median age for the control group in the present study was 24.3 years. It is felt that, in general, individuals with Turner's syndrome have as high an incidence of otitis media as the normal population.

Audiometric Analyses[11]

Definitions

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[9]V. Howie, "Acute and Recurrent Otitis Media," *Hearing Loss in Children*, ed. B. Jaffe (Baltimore: University Park Press, 1977), pp. 421-430.

[10]Paparella and Kuhn, *op. cit.*

[11]See Appendices A, B, and C.

Hearing was considered to be within normal limits if the threshold was ≤ 15 dB in the frequency range of .250-8.0 kHz. Hearing was considered to be impaired if the threshold was at least 20 dB or worse at one or more frequencies. *Acoustic reflex thresholds* were considered to be within normal limits if they occurred between 70-100 dB SL. When the reflex occurred at sensation levels of ≤ 60 dB, *recruitment* was inferred. *Middle ear function* was defined as normal when the ears exhibited a middle ear resting pressure of ±980 Pa. *Eustachian tube function* was considered normal when a pressure change of ≥ 49 Pa and a function change of ≥ 5 percent occurred.

Pure Tone Threshold

Of the Turner's subjects (58 ears), only 18 ears (31 percent) were found to have normal auditory sensitivity according to the above definition. Sensorineural impairments predominated (38 percent), but a large number were of the mixed type (24 percent). Pure conductive impairments were the least represented (5 percent). These results are somewhat inconsistent with the preliminary observations of Lindsten (1963) and Anderson, et al. (1969) who reported an incidence rate for mixed losses of 3-7 percent and for conductive losses of 10-25 percent. The number of sensorineural losses found in the present study is fairly consistent with the above authors' findings who reported an incidence rate for sensorineural losses in their populations of 49-53 percent.

1. Of the 22 ears with sensorineural impairments in this study, 15 ears had impairments in the higher frequencies (6.0-8.0 kHz) only and were of a magnitude of 20-50 dB HL. The other seven ears had

impairments across the frequencies.

2. The dip previous investigators referred to was found in nine ears. These dips were generally located between 1.0-6.0 kHz with a magnitude range of 20-45 dB and a mean value of 34 dB HL. The location and magnitude of this audiometric pattern was consistent with those found in the studies of Anderson, et al. (1969) and Lindsten (1963).

3. Mixed losses were found in 14 ears; in six of these the sensorineural component was located at 6.0-8.0 kHz only.

4. Four ears displayed pure conductive impairments, all of which were mild in degree. Three of these demonstrated a loss of 20 dB HL at one frequency while the remaining one covered a two frequency range.

5. In general, pure tone testing revealed a higher incidence of mixed impairments and a lower incidence of conductive impairments than was previously reported in the Turner's population.

These results lend support to the basin-shaped threshold curve Anderson, et al. (1969) found characteristic of the sensorineural losses in Turner's; however, these hearing impairments were not severe enough to pose communication difficulties for the population but do warrant monitoring due to the possible progressive nature found by Anderson, et al. It is felt that additional research is needed to more clearly describe the hearing impairments, resolve the discrepancies found between this study and others, as well as delineate the progressiveness of the hearing disorders in the Turner's population.

Acoustic Reflexes

In this investigation the acoustic reflex threshold was judged to be reduced when a span of 60 dB SL or less was found between the audiometric and reflex thresholds in at least one stimulus frequency. In contrast, a reflex threshold was judged to be elevated when it occurred at levels of 105 dB SL or greater in at least one frequency.

Table 1 indicates the results of the reflex testing across the various stimulus frequencies. Absent reflexes occurred more often than did any of the abnormal reflex conditions and occurred more often at the higher stimulus frequencies (2.0 kHz). Reflexes were absent in three ears across all frequencies. In two of these ears tympanometry indicated stiff middle ear systems due to the presence of fluid. In the third ear, middle ear function and auditory thresholds were normal. An additional two ears also displayed absent reflexes, but these absences occurred at only one frequency. They, too, evidenced normal middle ear function and normal auditory sensitivity.

Failure to elicit a reflex may be due to several factors (Wilber, 1976):

1. Middle ear pathology.
2. Nerve VIII damage of the stimulated ear.
3. Severe hearing loss in the stimulated ear.
4. Absence of a stapedial tendon in the stimulated ear.
5. Nerve VII damage in the nontest ear.
6. Inability of the equipment to produce a signal intense enough to elicit a reflex.

Table 1

Occurrence of the Acoustic Reflex as it Relates to Probe Tone Frequency and Reflex-eliciting Frequency

Acoustic reflexes	Turner's group					Control group				
	220 Hz			660 Hz		220 Hz			660 Hz	
	.500 kHz	1 kHz	2 kHz	1 kHz	2 kHz	.5 kHz	1 kHz	2 kHz	1 kHz	2 kHz
Normal	16*	17	14	19	19	19	20	17	20	18
Elevated	3	1	1	2	2	11	0	3	0	2
Reduced	0	1	2	3	4	0	0	0	0	0
Absent	3	3	5	3	4	0	0	0	0	0

*Ears.

The first two factors may be ruled out based on the testing completed although no additional testing was conducted to rule out the other factors.

Some investigators suggest that elevated reflex thresholds could be indicative of neural pathology (Jerger and Jerger, 1974; Olsen, et al., 1975). In the Turner's group, elevated reflex thresholds were found across all stimulus frequencies with both probe tone frequencies. A larger percentage of ears showed elevated reflexes at 500 Hz (14 percent) with the percentage of occurrence across the remaining frequencies ranging from 5-7 percent (see Table 1).

Of the abnormal reflex states only elevated reflex thresholds were found to occur in the control group. Utilizing the 220 Hz probe tone frequency, one ear was found to have an elevated reflex threshold at 500 Hz and three ears had elevated thresholds at 2.0 kHz. With the 660 Hz probe tone, two ears demonstrated elevated reflex thresholds at 2.0 kHz. It appears that in the control group the stimulus frequency of 2.0 kHz is more sensitive to elevated reflexes than the lower stimulus frequencies.

Reduced reflex thresholds indicative of recruitment (to some investigators) (Anderson, et al., 1969; Jerger, et al., 1972), were found at the higher stimulus frequencies (1.0-2.0 kHz); they were also found more often with the higher probe tone frequency in the Turner's group. One ear (4 percent) was found to have reduced reflex thresholds at 1.0 kHz and two ears (9 percent) had reduced thresholds at 2.0 kHz with the 220 Hz probe frequency. Utilizing the 660 Hz probe tone, 11 percent and 14 percent of the ears had elevated reflexes at 1.0 kHz and

2.0 kHz, respectively. It would appear that, given the parameter used in this study to indicate a reduced span getween the audiometric and reflex threshold (105 dB SL or greater), the 660 Hz probe tone frequency is more sensitive in indicating the reduced range than the lower probe frequency.

This author found the acoustic reflex thresholds of the Turner's group to be consistent with the types of hearing impairments displayed by the individuals. Reflex thresholds were found to be absent in four ears with conductive components, reduced in five ears with sensorineural losses, and elevated in three ears with normal thresholds.

Eustachian Tube Function

Of the 11 Turner's subjects on whom the pressure swallow test was performed, seven subjects exhibited middle ear pressure changes of greater than 49 Pa and middle ear function changes of greater than 5 percent, indicative of functioning eustachian tubes.

As a group, the Turner's subjects did not differ significantly from the control group or the normal population. Comparing results with those obtained by Seidemann and Givens (1977) in their study conducted with 51 normal children (mean age = 7.6 years), it was found that 64 percent of the Turner's subjects and 56 percent of the Seidemann and Givens' subjects displayed pressure/function changes; however, when pressure changes and function changes are examined, the Turner's subjects obtained significantly more changes than the normal children (see Table 2).

Comparing results with those obtained by Seifert, et al. (1979),

Table 2

Comparison of Middle Ear Measurement Change Following Eustachian Tube Test

Middle ear change	No. of ears	%	$\bar{X}$
TURNER'S GROUP*			
Pressure only	17	77	24.54
Function only	19	86	17.41
Pressure and function	14	64	-
No change	0	0	-
CONTROL GROUP*			
Pressure only	20	91	21.59
Function only	19	86	17.27
Pressure and function	18	82	-
No change	1	5	-
SEIDEMANN AND GIVENS†			
Pressure only	2	2.3	-
Function only	26	29.2	-
Pressure and function	56	62.9	-
No change	5	5.6	-

Middle ear change	S.D.	$\bar{X}$
SEIFERT, ET AL.‡		
Pressure change	12.60	13.5
Function change	15.15	22.0

*Present study, 22 ears.

†Fifty-one children, 89 ears.

‡Twenty-six adults, 48 ears.

who examined 26 normal adults (mean age = 23.6), it was found that both the Turner's group and the control group were within one standard deviation from the mean of the adults. No significant differences were found.

Examination of data of the individual ears leads one to question the validity of the pressure swallow test as an indicator of tubal function. According to the procedures outlined in this study, a eustachian tube was said to be hypofunctioning when a middle ear pressure change of less than 49 Pa and a middle ear function change of less than 5 percent were obtained. The literatures states (Feldman and Wilber, 1976; Seifert, et al., 1979; Williams, 1975) that there are other factors which may also suggest a hypofunctioning tube: an abnormal negative middle ear pressure or change in pressure in a negative direction on the pressure swallow test.

Of the 14 ears which displayed pressure/function changes indicating normal tubal function, six ears demonstrated pressure changes in a negative direction and one ear demonstrated a middle ear pressure of -2060 Pa. Three ears had mild conductive impairments and three ears had absent acoustic reflexes--other conditions which may influence eustachian tube function. One would expect to find, in these instances, some evidence of a hypofunctioning eustachian tube.

In an examination of the eight ears with nonfunctioning eustachian tubes, one finds four ears with normal auditory thresholds and one ear with a sensorineural impairment throughout the frequency range. One might expect to find functioning eustachian tubes in these cases.

The Turner's subjects did not, as a group, vary from the control group or the normal population on the pressure swallow test. Examination of the individual data, however, reveals contradictory data leading one to question the validity of this procedure. Functioning eustachian tubes were found in individuals with abnormal middle ear pressures, and conductive losses and nonfunctioning eustachian tubes were found in individuals with normal auditory thresholds and normal middle ear pressures.

Chapter 5

SUMMARY AND CONCLUSIONS

Summary

The results of the present study indicate that the majority of the Turner's subjects did have an auditory impairment. A sensorineural hearing loss affecting 6.0 and 8.0 kHz only, was the most common. A large number of mixed impairments were also found with a large majority of the sensorineural components occurring at 6.0 and 8.0 kHz.

While the Turner's subjects had as high an occurrence of otitis media as the normal population, a large majority of them had middle ear infections after the age puberty normally begins and a few continue to have the infections. Some investigators (English, et al., 1973; Paparella and Brady, 1970) believe that sensorineural impairments can be a natural sequela of chronic otitis media. This leads one to speculate about the large number of high frequency losses found in this study. Could they, too, be the result of otitis?

Examination of acoustic reflexes revealed a consistency with the pure tone configuration in most cases. Reflexes were within normal limits in subjects with normal hearing sensitivity and reduced in some subjects with sensorineural impairments.

The eustachian tube function test, used to predict tubal functioning, resulted in contradictory data. One subject displayed abnormal middle ear pressure, an indication of tubal dysfunction, but the pressure swallow test indicated a normal functioning eustachian tube. The opposite was also found to be true.

There is some question about using a middle ear pressure change of 49 Pa as one parameter of an adequately functioning eustachian tube. According to Williams (Feldman and Wilber, 1976) instrument hysteresis will result in changes of similar magnitude, 39.2-88.2 Pa. Too, since middle ear tissues absorb approximately 490 Pa an hour (Elner, 1977) in six minutes, approximately the length of time it takes to complete the pressure swallow test on one ear, a 49 Pa change will occur by normal processes alone.

There is also a question about the validity of using a 5 percent change in middle ear function as another parameter of eustachian tube function. Examination of the acoustic-immitance research (Margolis and Popelka, 1977) reveals a much more complex interaction among the tympanometric variables which, in reality, one is measuring with the pressure swallow test. The procedure uses a 5 percent change in conductance (GA) to indicate tubal function; however, a 5 percent change in conductance is not equal to a 5 percent change in impedance (Z), the indicator of middle ear function. Until further research has been completed with the pressure swallow test, one should question its validity as an indicator of eustachian tube function.

Conclusions

As a pilot project, this study has raised more questions than it has answered. Hopefully, some of these questions can be answered by a longitudinal audiological survey of the Turner's population.

1. Do Turner's individuals have otitis media longer than the normal population and, if so, may the infections be the cause of the high frequency and sensorineural impairments that are found?

2. What types of auditory impairments do the majority of the Turner's population have--conductive, sensorineural, high frequency losses--and are their losses progressive?

3. Why was there such a high incidence of elevated reflexes in the two populations studied? Is this typical of the populations in general?

4. Would a eustachian tube function test demonstrate differences in tubal function between the Turner's population and the normal population and, if so, is it often enough to warrant inclusion in a diagnostic battery?

BIBLIOGRAPHY

American National Standards Institute. Guidelines for Acoustic-immitance Measurements, 1978.

American National Standards Institute. American National Standards Specifications for Audiometers, S3.6-1969.

Anderson, H., R. Filipsson, E. Fleur, B. Kock, J. Lindsten, and E. Wedenberg. "Hearing Impairment in Turner's Syndrome." *Acta-Otolaryngologica* Supp. 247, 1969.

Bluestone, C., J. Paradise, and Q. Berry. "Physiology of the Eustachian Tube in the Pathogenesis of Middle Ear Effusions." *Laryngoscope,* 82(1972), 1654-1670.

Carhart, R. and J. Jerger. "Preferred Method for Clinical Determination of Pure Tone Thresholds." *Journal of Speech and Hearing Disorders,* 24(1959), 330-345.

Elner, A. "Quantitative Studies of Gas Absorption from the Normal Middle Ear." *Acta-Otolaryngologica,* 83(1977), 25-28.

Engel, E. and A. Forbes. "Cytogenetic and Clinical Findings in 48 Patients with Congenitally Defective or Absent Ovaries." *Medicine,* 44(1965), 135-164.

English, G., J. Northern, and T. Fria. "Chronic Otitis Media as a Cause of Sensorineural Hearing Loss." *Archives of Otolaryngology,* 98(1973), 18-22.

Feldman, A. and L. Wilber, eds. *Acoustic Impedance and Admittance--the Measurement of Middle Ear Function.* Baltimore: Williams and Wilkins Co., 1976.

Ferguson-Smith, M. and others. "Clinical and Cytogenetical Studies in Female Gonadal Dysgenesis and Their Bearing on the Cause of Turner's Syndrome." *Cytogenetics,* 3(1964), 355-383.

Holmquist, J. "Eustachian Tube Function Assessed with Tympanometry." *Acta-Otolaryngologica,* 68(1969), 501-508.

Jaffe, B., ed. *Hearing Loss in Children.* Baltimore: University Park Press, 1977.

Jerger, J. "Clinical Experience with Impedance Audiometry." *Archives of Otolaryngology*, 92(1970), 311-324.

Jerger, J., ed. *Handbook of Clinical Impedance Audiometry*. Dobbs Ferry, New York: American Electromedics, 1975.

Jerger, J., D. Jerger, and L. Maudlin. "Studies in Impedance Audiometry: I. Normal and Sensorineural Ears." *Archives of Otolaryngology*, 96(1972), 513-523.

Jerger, J. and S. Jerger. "Audiological Comparison of Cochlear and Eighth Nerve Disorders." *Annals of Otolaryngology, Rhinnology, and Laryngology*, 83(1974), 275-286.

Jones, H., H. Turner, and M. Ferguson-Smith. "Turner's Syndrome and Phenotype." *The Lancet*, 1(1966), 1155.

Lindsten, J. *The Nature and Origin of X Chromosome Aberrations in Turner's Syndrome*. Stockholm: Almqvist, Wilsell, 1963.

Margolis, R. and G. Popelka. "Interactions Among Tympanometric Variables." *Journal of Speech and Hearing Research*, 20(1977), 447-462.

Martin, F., ed. *Pediatric Audiology*. New Jersey: Prentice-Hall, Inc., 1978.

Metz, O. "The Acoustic Impedance Measure on Normal and Pathological Ears." *Acta-Otolaryngologica* Supp. 63, 1946.

Olsen, W., D. Noffsinger, and S. Kurdziel. "Acoustic Reflex and Reflex Decay." *Archives of Otolaryngology*, 101(1975), 622-625.

Paparella, M. and B. Brady. "Sensorineural Hearing Loss in Chronic Otitis Media and Mastoiditis." *Archives of Otolaryngology*, 74 (1970), 108-115.

Rashad, M. and W. Morton, eds. *Genital Anomalies and Related Subjects*. Springfield: Charles Thomas, 1969.

Seidemann, M. and G. Givens. "Tympanometric Assessment of Eustachian Tube Patency in Children." *Journal of Speech and Hearing Disorders*, 44(1977), 487-497.

Seifert, M., M. Seidemann, and G. Givens. "An Examination of Variables Involved in Tympanometric Assessment of Eustachian Tube Function in Adults." *Journal of Speech and Hearing Disorders*, 44(1979), 388-395.

Silver, H. and S. Dodd. "Gonadal Dysgenesis." *AMA Journal of Diseases of Children*, 94(1957), 702-707.

Smith, D., ed. *Recognizable Patterns of Human Malformation.* Philadelphia: W. B. Saunders Co., 1976.

Stratton, H. "Gonadal Dysgenesis and the Ears." *Journal of Laryngology and Otology,* 79(1965), 343.

Szmeja, A. and J. Kosowicz. "Audiometric Studies in Turner's Syndrome." *Polish Endocrinology,* 18(1967), 300-303.

Szpunar, J. and M. Rybak. "Middle Ear Disease in Turner's Syndrome." *Archives of Otolaryngology,* 87(1968), 34-40.

Turner, H. "A Syndrome of Infantilism, Congenital Webbed Neck, and Cubitus Valgus." *Endocrinology,* 23(1938), 566-574.

Valkov, I., S. Dokumov, P. Genkova, and D. Dimov. "Olfactory, Auditory, and Gustatory Function in Patients with Gonadal Dysgenesis." *Obstetrics and Gynecology,* 46(1975), 417-418.

Wiet, R. and S. Coulthard, eds. *Proceedings of the Second National Conference on Otitis Media.* Columbus, Ohio: Ross Laboratories, 1978.

Williams, P. "A Tympanometric Pressure Swallow Test for Assessment of Eustachian Tube Function." *Annals of Otolaryngology,* 84(1975), 331-343.

Wolstenholme, G. and J. Knight, eds. *Sensorineural Hearing Loss.* London: J. A. Churchill, 1970.

APPENDIX A

Audiometric Results

Case no.	Init-ials	Age (yrs)	Tympanometry*		Acoustic reflexes†		E. T. test‡		Type of impairment**
			R	L	220 Hz	660 Hz	R	L	
1	SY	19.0	N	N	N	N	-	-	Right: cond., 20 dB @ 250 Hz Left: ----
2	MH	13.0	N	N	A	A	-	-	Right: cond. with HF loss 30 dB @ 250 Hz; 20 dB @ 8 kHz Left: HF = 25 dB @ 6 k, 46 dB @ 8 k
3	PR	10.5	N	N	E	N	-	+	Bil. cond. with dip Right: 20 @ 250 Hz; dip at 6 kHz = 30 dB Left: 20 @ 250 Hz; 35 dB @ 6 k and 8 kHz
4	AS	18.42	N	Hypermobile	DNT	DNT	-	+	Right. ---- Left: ----
5	JB	21.42	N	N	N	E	+	+	Bil., sym., HF, dip 6 kHz = 25-30 dB
6	CP	17.75	N	-105 mm	N	N	-	+	Right: ---- Left: cond.; 20 @ 250 Hz
7	JP	13.25	N	-210 mm	E	E	+	-	Right: ---- Left: ----
8	RC	18.66	N	N	N	N	+	+	Bil., sens. Right: dip 3-6 kHz; Max. 6 kHz = 25 dB Left: flat, 4-8 kHz = 30 dB

(continued next page)

Case no.	Init-ials	Age (yrs)	Tympanometry*		Acoustic reflexes†		E. T. test‡		Type of impairment**
			R	L	220 Hz	660 Hz	R	L	
9	SM	23.17	N	N	E	E	-	+	Right: cond., HF; 20 dB @ 250 Hz, 20 @ 6 kHz Left: ----
10	TE	14.33	F	F	A	A	CNT	CNT	Bil., cond. Right: 35-40 dB Left: 25 dB @ 250, 20 dB @ 6-8 kHz
11	SH	13.33	N	N	D	N	+	+	Right: sens., dip 1-4 kHz, Max. 2-3 kHz of 50 dB
12	CM	40.58	N	N	D	D	-	+	Bil., sens. Right: sloping 3-8 kHz = 20-55 dB Left: dip 2-8 kHz, Max 3 kHz = 45 dB
13	TM	17.42	P	S	DNT	DNT	DNT	DNT	Bil., cond. Right: upward sloping .250-1 kHz = 45-10 dB, flat 1.5-8 kHz = 35-45 dB
14	CM	26.33	N	N	N	D	DNT	DNT	Bil., sens. Right: flat 1-8 kHz = 20-35 dB Left: flat 25-45 dB
15	JT	16.33	N	N	N	N	DNT	DNT	Right: ---- Left: 30 @ 8 kHz
16	MB	11.58	-	-	DNT	DNT	DNT	DNT	Right: HF, 30-40 dB @ 6-8 kHz Left: cond., HF, 25 dB @ 250 Hz, 20-40 dB @ 4-8 kHz

(continued next page)

Case no.	Init- ials	Age (yrs)	Tympanometry*		Acoustic reflexes†		E. T. test‡		Type of impairment**
			R	L	220 Hz	660 Hz	R	L	
17	AF	14.08	N	N	DNT	DNT	DNT	DNT	Bil., HF Right: 35-50 dB @ 6-8 kHz Left: 20-25 @ 6-8 kHz
18	VP	9.58	N	N	DNT	DNT	DNT	DNT	Right: ---- Left: ----
19	CL	12.17	N	N	DNT	DNT	DNT	DNT	Right: ---- Left: 30 dB @ 1 kHz
20	AG	11.33	N	N	DNT	DNT	DNT	DNT	Bil., HF Right. 20 dB @ 8 kHz Left: 20 dB @ 8 kHz
21	CW	17.0	N	N	DNT	N	DNT	DNT	Bil., HF Right: 20-25 dB @ 6-8 kHz Left· dip 6-8 kHz, Max. 6 kHz = 35 dB
22	KC	13.08	N	N	DNT	DNT	DNT	DNT	Bil., HF Right· dip 4-8 kHz, Max. 4 kHz = 45 dB Left: 25 dB @ 8 kHz
23	VC	9.5	PC	Tubes	DNT	DNT	DNT	DNT	Bil., cond., HF Right: 35 dB @ 240 Hz; 20 dB @ 2-8 kHz Left: rising, 35-25 dB @ 250-1 kHz; 20 dB @ 8 kHz

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Case no.	Init-ials	Age (yrs)	Tympanometry*		Acoustic reflexes†		E. T. test‡		Type of impairment**
			R	L	220 Hz	660 Hz	R	L	
24	JW	15.33	N	N	DNT	DNT	DNT	DNT	Right: ---- Left: ----
25	KV	17.92	-	-	DNT	DNT	DNT	DNT	Bil., cond., HF Right· rising 15-50 dB @ 1250-2 kHz; sloping 25-65 dB @ 3-8 kHz Left: flat 20-25 dB @ .250-1 kHz, sloping 20-60 dB @ 4-8 kHz
26	JR	14.08	N	N	DNT	N	DNT	DNT	Right: ---- Left. 20 dB @ 8 kHz
27	MY	15.08	-	-	DNT	DNT	DNT	DNT	Right: HF, 25 dB @ 8 kHz Left: cond., 20 dB @ 250-500 Hz
28	SS	18.08	N	N	DNT	DNT	DNT	DNT	Right: HF, dip, 6-8 kHz, Max. 6 kHz = 25 dB Left: ----
29	SS	13.42	N	N	N	N	DNT	DNT	Right: ---- Left: ----

*N = normal tracing
F = flat tracing with no point of maximum compliance
P = perforation
S = stiff middle ear system
-105 mm, -210 mm - abnormal negative middle ear pressure in milimeters of H_2O
Hypermobile = large peak amplitude at atmospheris pressure

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†N = normal acoustic reflex thresholds (see text for explanation)
A = absent acoustic reflex thresholds
E = elevated acoustic reflex thresholds
D = depressed acoustic reflex thresholds
DNT = did not test

‡E. T. test = eustachian tube function test (see text for explanation)
+ = functioning eustachina tube
- = hypofunctioning eustachian tube
CNT = could not test

** ---- = within normal limits
cond. = conductive hearing impairment
sens. = hearing loss of pure sensorineural origin
HF = high frequency hearing loss
Max. = maximum
bil. = bilateral
sym. = symmetrical, 10 dB difference between ears
dB = decibel above normal threshold of hearing
kHz = test tone frequency in cycles per second divided by 1,000
dip = basin-shaped threshold curve; figures following denote frequency range of basin extent
flat = threshold curve that is reasonably horizontal
rising = threshold curve gradually rising toward higher frequencies (dB figures denote beginning and maximum of hearing loss range)
sloping = threshold curve gradually falling toward higher frequencies

APPENDIX B

Acoustic Reflex Thresholds in dB SL for the Turner's Group

Case no.	Initials	Age (yrs)	Acoustic reflex thresholds									
			220 Hz probe tone						660 Hz probe tone			
			500 Hz		1,000 Hz		2,000 Hz		1,000 Hz		2,000 Hz	
			R	L	R	L	R	L	R	L	R	L
1	SY	19.0	75	90	90	85	95	-*	90	85	95	90
2	MH	13.0	-	90	-	95	-	-	-	90	-	90
3	PR	10.5	95	80	85	90	90	85	85	90	90	90
5	JB	21.42	90	90	85	85	90	80	95	85	100	80
6	CP	17.75	90	95	90	95	95	80	90	95	90	85
7	JP	18.86	110	105	100	90	-	85	100	90	-	90
8	RC	18.66	90	95	90	85	85	90	85	80	80	85
9	SM	23.17	-	105	110	100	105	90	110	105	105	105
10	TE	14.33	75	-	-	-	-	-	-	-	-	-
11	SH	13.33	85	85	70	80	55	85	60	75	60	85
12	CM	40.58	70	70	70	60	65	50	70	65	60	50
14	CM	26.33	DNT†	DNT	DNT	DNT	DNT	DNT	55	55	60	65

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Case no.	Initials	Age (yrs)	Acoustic reflex thresholds									
			220 Hz probe tone						660 Hz probe tone			
			500 Hz		1,000 Hz		2,000 Hz		1,000 Hz		2,000 Hz	
			R	L	R	L	R	L	R	L	R	L
15	JT	16.33	DNT	DNT	DNT	DNT	DNT	DNT	80	85	85	85
21	CW	17.0	DNT	DNT	DNT	DNT	DNT	DNT	DNT	85	DNT	85
26	JR	14.08	DNT	DNT	DNT	DNT	DNT	DNT	DNT	DNT	90	80
29	SS	13.42	95	90	90	90	90	90	90	85	90	90

*- = no response

†DNT = did not test

A P P E N D I X   C

Measure of Eustachian Tube Function of the Turner's Group

Case no.	Initials	Eustachian tube function test: Function change (%) R	L	Pressure change (Pa) R	L
1	SY	15	-3	0	245
2	MH	18	21	-49	588
3	PR	12	31	0	1079
4	AS	-4	24	1177	-1079
5	JB	42	11	-441	392
6	CP	12	10	0	-49
7	JP	20	16	49	0
8	RC	18	23	49	245
9	SM	17	8	0	-441
11	SH	23	17	196	49